THORACOSCOPIC ABLATION IN SITUS INVERSUS DEXTROCARDIA WITH INTERRUPTED INFERIOR VENA CAVA CONTINUATION IN AZYGOS VEIN

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Abstract

INTRODUCTION Situs inversus totalis, dextrocardia with interrupted inferior vena cava and azygos vein continuation concomitant with symptomatic atrial fibrillation requiring ablation. This case was deemed not suitable for percutaneous ablation due to anatomic variations and the lack of case reports in literature. METHODS AND RESULTS We performed bilateral thoracoscopic epicardial ablation and epicardial left atrial appendage exclusion. The direct vision allowed for a complete box lesion set with bipolar radiofrequency device. Patient remained in sinus rhythm at 12-months follow-up. CONCLUSION Surgical thoracoscopic epicardial ablation is safe and effective also in congenital defects. Multidisciplinary expertise can offer minimally invasive ablation treatments.

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METHODS AND RESULTS

We performed bilateral thoracoscopic epicardial ablation and epicardial left atrial appendage exclusion. The direct vision allowed for a complete box lesion set with bipolar radiofrequency device. Patient remained in sinus rhythm at 12-months follow-up.

CONCLUSION

Surgical thoracoscopic epicardial ablation is safe and effective also in congenital defects. Multidisciplinary expertise can offer minimally invasive ablation treatments.

KEYWORDS

Atrial fibrillation, radiofrequency ablation, surgical thoracoscopic ablation, minimally invasive, left atrial appendage, left atrial appendage exclusion, situs inversus totalis, dextrocardia, interrupted inferior vena cava

INTRODUCTION Concomitant long-persistent atrial fibrillation in a rare congenital aberration: situs inversus totalis, dextrocardia with interrupted inferior vena cava and azygos vein continuation. This case was deemed not suitable for percutaneous ablation due to anatomic variations and the lack of case reports in literature. METHODS AND RESULTS We performed surgical thoracoscopic epicardial ablation and concomitant left atrial appendage exclusion. Thanks to direct vision, the complete box lesion was performed with radiofrequency. At twelve months patient remained in sinus rhythm. CONCLUSION Surgical thoracoscopic epicardial ablation is safe and effective also in congenital defects. Multidisciplinary expertise can offer minimally invasive ablation treatments.

KEYWORDS: Atrial fibrillation, radiofrequency ablation, surgical thoracoscopic ablation, left atrial appendage, left atrial appendage exclusion, congenital defects, situs inversus totalis, dextrocardia, interrupted inferior vena cava

INTRODUCTION

Intrahepatic interruption of the inferior vena cava (IVC) with azygos vein continuation is a rare congenital aberration, occurring in 0.6% of patients with other cardiac defects as situs abnormalities and dextrocardia.[1] In most instances, patients are asymptomatic. They might be noticed as incidental findings following imaging investigations. We present a case of situs inversus totalis and dextrocardia associated with interrupted inferior vena cava (IVC) and azygos vein continuation in a patient with concomitant long-persistent atrial fibrillation. The treatment of atrial fibrillation (AF) has been considered at high risk for percutaneous interventions and thus referred to our institute for surgical bilateral thoracoscopic epicardial ablation and left atrial appendage (LAA) exclusion.

The patient is a 45-years old male with symptomatic long-standing persistent atrial fibrillation (LsP-AF) who received multiple electrical cardioversions for AF recurrences since 2003. Class I antiarrhythmic drugs failed to restore sinus rhythm thus, according to the current ESC 2020 Guidelines for AF treatment [2], the patient was scheduled for transcatheter pulmonary veins (PV) isolation.[3]Pre-operative chest X-RAY showed the presence of a complete situs inversus dextrocardia (SID). (Figure 1A)

Then, a thoraco-abdominal CT scan was performed to rule out other anatomical abnormalities. A concomitant intrahepatic interruption of the IVC was described with the renal veins draining into the azygos vein which was directly collecting blood from the lower body and draining posteriorly into the superior vena cava

(SVC). In adjunct, the hepatic veins were draining directly into the right atrium (RA). Nonetheless, regular anatomy of the left atrium (LA) with two right and two left PVs was depicted (Figure 1B, 1C).

The patient was deemed not suitable for percutaneous ablation (PA) because of the complex anatomy and was then referred to our Institution for surgical thoracoscopic ablation. Transesophageal echocardiogram was performed to rule out thrombus in the LAA prior to surgery.

METHODS

Thoracoscopic procedure was performed starting from the chest side in which LAA was promptly approachable: the right side. Two 10-mm ports and one 5-mm port were placed along the right anterior axillary (3th and 5th intercostal space) and mid-axillary line (4th intercostal space) as previously described for classic anatomy procedures.[3,4] Briefly, once the pericardium was opened, the LAA was visualised as reference. A blunt dissection at the level of the roof of the LA was performed to allow the right curved bipolar ablation device (Isolator Synergy Clamp, EMR2; AtriCure Inc, Mason, OH USA) to encircle the PV on the right side (Figure 2A). The device followed the route previously prepared by means of a track-light dissector (Lumitip, AtriCure Inc, Mason, OH USA). Connecting lesions were then performed by using a linear bipolar radiofrequency device (Coolrail linear pen, AtriCure Inc, Mason, OH USA) at the level of the inferior side of the Box Lesion (Figure 2B). LAA occlusion was performed by means of AtriClip Pro 2 device 40 mm (AtriCure Inc, Mason, OH USA). We then moved ports to the contralateral side in order to complete the thoracoscopic surgical ablation (Figure 2C).

A blunt dissection of the pericardial reflection below the confluence of the two sovra-hepatic veins (draining into the RA) was performed in order to gain access to the oblique sinus (Figure 3A). The access to the transverse sinus was obtained by gently dissecting the pericardial reflection below the SVC. PVs at the left side were encircled and RF energy was delivered by means of left curved bipolar ablation RF device (Isolator Synergy Clamp, EML2 AtriCure Inc, Mason, OH USA) (Figure 3B). Once completed, connecting lesions between right and left PVs were performed at the level of the roof of the LA (Coolrail, AtriCure Inc, Mason, OH USA). Exit block was confirmed at the end of the procedure with a total surgical time of 200 minutes.

RESULTS Intra-operatively and during hospitalization no complications occurred. The patient was discharged in sinus rhythm after four days. At three and twelve months follow-up, the patient was in stable sinus rhythm without experiencing recurrent symptomatic episodes of AF, as confirmed by means of holter-ECG evaluation.

DISCUSSION

The incidence of dextrocardia with situs inversus totalis is 1/10,0000–50,000 births.(6) In such patients, the IVC might be interrupted or stenotic in 8-18% of patients, with azygos continuation present in only 0,6% of cases.[7] Unless severe concomitant congenital defects occur, patients with SID showed life expectancy similar to that of the general population.[8] In literature only few case reports described ablation strategies in this specific subset of patients, in which percutaneous PVs isolation can be extremely challenging. The major limitations are considered: small calibre of the entrance vessel, often complicated by tortuosity especially at the level of the azygos vein;[9] difficulty in obtaining trans-septal puncture; limited manoeuvrability of the ablation/mapping system.[7] For these reasons, "unconventional methods" were explored and described in literature:[10,11] so far, three different PA approaches have been reported: trans-septal puncture via transjugular approach through the SVC[9]; transaortic retrograde approach[10] and percutaneous transhepatic vein approach.[11]

Trans-septal puncture via internal jugular/subclavian vein and SVC was described by Masumoto et al.[9] PVs isolation was effectively performed under 3D navigation system. However, the procedure was reported as extremely long despite authors simplified the procedure by omitting electrophysiological mapping of the LA (over 360 minutes of procedural time with more than 60 minutes of PVI time). Major concerns raised mainly while performing the trans-septal puncture with a procedure generally defined as "not smooth". Of note, authors avoided to approaching the IVC because of the tortuosity.[9]

An intriguing trans-aortic solution was reported by Okajima and colleagues [10] in this specific subset of patients. Via the left femoral artery authors gained access to the LA in a retrograde fashion and under magnetic navigation system. However, authors were unable to perform a complete isolation of the four PVs: the right inferior PV isolation was aborted since the ablation catheter repeatedly dropped into the left ventricle because of an unfavourable angle and short distance between mitral annulus and PV orifice. Moreover, in order to simplify a such complex procedure, authors avoided to evaluate entrance/exit block as well as the electro-anatomical mapping. [9,10]

A percutaneous trans-hepatic vein approach was described by Tandon et al.[11] in a patient with dextrocardia and IVC continuation in the azygos vein. LA access was gained via hepatic vein puncture under ultrasound guidance and fluoroscopy. The procedure was reported as safe, however, only addressed the LAA. [11]

Non-invasive percutaneous solutions provided a stable restoration of the sinus rhythm, however they were reported as particularly challenging. In most instances, the major drawbacks were the necessity to use different types of catheters, the complex catheter guidance and manoeuvrability, thus leading to exceedingly long and often incomplete procedures.

Surgical thoracoscopic AF ablation had the advantage of a direct vision of the complex anatomy of the patient, allowing the surgeons to promptly recognize cardiac structures despite the complete SID. Effective lesions were safely performed and confirmed by testing the presence of the exit block, thus without affecting the completeness of the ablation lesion set and the procedural time. Moreover, the LAA was successfully excluded by epicardial access with no additional risk.

CONCLUSION In conclusion, patients with complex anatomy should be carefully evaluated with a multidisciplinary approach in specialistic facilities with a wide expertise in minimally invasive arrhythmia surgery. A patient-tailored approach was guaranteed once risks and benefits of the surgical procedure over percutaneous strategy were wisely balanced, thus providing the best option in terms of safety, efficacy and patient satisfaction.

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FIGURE LEGEND

Figure 1 - A: chest X-Ray showing complete situs inversus dextrocardia. B: Left-sided view howing azygos vein (white asterisk) continuation draining into the posterior aspect of the superior vena cava (white dot). C: Posterior view of the azygos vein (white asterisk) crossing the midline.

Figure 2 – A: Right curved bipolar ablation device encircling the right-sided pulmonary veins (white arrow: right superior pulmonary vein; white asterisk: left atrial appendage). B: Linear radiofrequency device performing connecting lesion at the level of the floor of the box lesion (white arrow: left inferior pulmonary vein; white asterisk: confluence of the hepatic veins). C: Left atrial appendage exclusion device positioning.

Figure 3 – A: Left anatomy after pericardial opening. B: Left curved bipolar ablation device encircling the left sided pulmonary vein. (white arrrow: left pulmonary vein; white asterisk: superior vena cava).